

Portal hypertension complicating myelofibrosis in a patient without portal or hepatic vein thrombosis

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In the setting of myelofibrosis, portal hypertension often results from portal and/or hepatic vein thrombosis. However, intrahepatic non-cirrhotic portal hypertension without portal or hepatic vein thrombosis is rarely described in these patients [1,2]. Herein, we report a case of myelofibrosis presenting with variceal bleeding in the absence of portal vein or hepatic vein thrombosis.

A 23-year-old female patient presented with massive hematemesis and melena at the emergency department. She had no history of viral hepatitis, alcohol abuse, or any other liver diseases, but she had been diagnosed with JAK^{V617F}

mutation (i.e., the V617F mutation of the Janus kinase 2 gene) negative myelofibrosis by bone marrow biopsy three years ago. Physical examination revealed that the spleen was palpable about 20 cm below the left costal margin. Laboratory tests showed a decreased platelet count of $17 \times 10^9/L$ and hemoglobin concentration of 70 g/L, a relatively normal liver function (alanine aminotransferase: 8 U/L, total bilirubin: 18.7 μ mol/L, albumin: 30.4 g/L) and prothrombin time of 15.5 sec. Hepatitis B virus surface antigen and anti-hepatitis C virus antibody were negative. Abdominal color Doppler ultrasound and contrast-enhanced computed tomography demonstrated the patent portal vein and hepatic veins, massive splenomegaly, and no ascites. Upper gastrointestinal endoscopy confirmed that upper gastrointestinal bleeding was caused by large esophageal varices. Liver biopsy was not performed due to a significantly low platelet count. Thus, she was diagnosed with portal hypertension secondary to myelofibrosis. Blood transfusion, octreotide, and prophylactic antibiotics were performed at the emergency department. After that, bleeding was controlled. This patient was transferred to our department of Digestive Interventional Radiology. At this time, laboratory tests demonstrated that hemoglobin concentration was elevated to 83 g/L with a relatively normal liver function (alanine aminotransferase: 16 U/L, total bilirubin: 11.3 μ mol/L, albumin: 33.2 g/L) and prothrombin time of 14.7 sec. A transjugular intrahepatic portosystemic shunt was planned, but the patient did not consent. Thus, propranolol was prescribed for the prevention of variceal rebleeding. She was doing well without any episodes of variceal rebleeding about one year after discharge.

Comparably with previous case reports, our case suggests the possibility of intrahepatic non-cirrhotic portal hypertension without portal or hepatic vein thrombosis in myelofibrosis. In clinical practice, myelofibrosis should be excluded in patients with symptomatic portal hypertension of unknown cause. However, due to the absence of liver biopsy, we are uncertain about the presence of liver infiltration from myeloid cells.

References

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